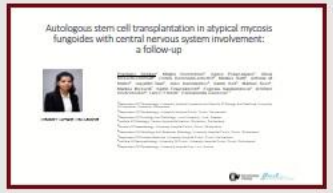
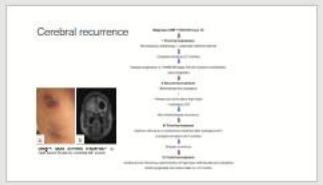
Pause 00:00:00 Select Area Audio Record Pointer

New Slide Table Pictures Screenshot Photo Album Shapes SmartArt Chart Get Add-ins My Add-ins Link Action Comment Text Box Header & Footer WordArt Date & Time Slide Number Object Equation Symbol Video Audio Screen Recording

Slides Tables Images Illustrations Add-ins Links Comments Text Symbols Media





Autologous stem cell transplantation in atypical mycosis fungoides with central nervous system involvement: a follow-up

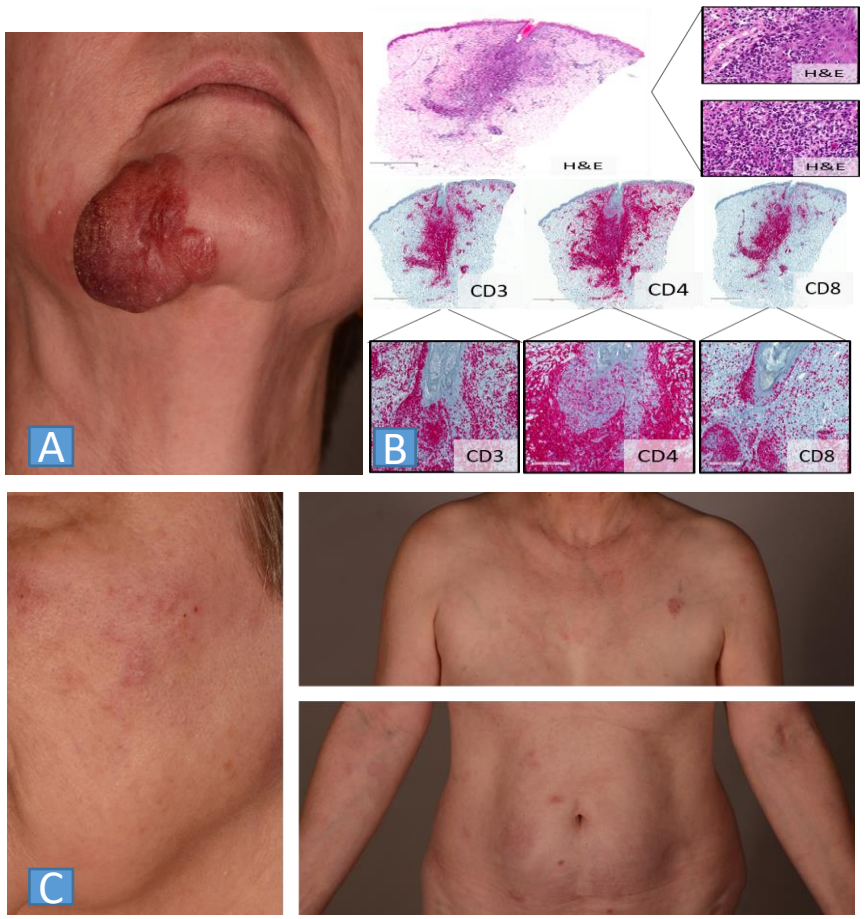
Prachitee Sirsikar, PhD Student

Prachitee Sirsikar[1], Mirjam Doerschner[2], Agnes Pekar-lukacs[3], Olivia Messerli-odermatt[2], Corina Dommann-scherrer[4], Markus Rütti[5], Antonia M Müller[6], Gayathri Nair[7], Jivko Kamarachev[8], Katrin Kerl[2], Markus Beer[6], Markus Messerli[7], Katrin Frauenknecht[8], Eugenia Haralambieva[6], Wolfram Hoetzenecker[9], Lars E French[2], Emmanuella Guenova[1,2]

[1]Department Of Dermatology, University Hospital Lausanne And Faculty Of Biology And Medicine, University Of Lausanne, Lausanne, Switzerland.
[2]Department Of Dermatology, University Hospital Zurich, Zurich, Switzerland.
[3]Department Of Oncology And Pathology, Lund University, Lund, Sweden.
[4]Institute Of Pathology, Canton Hospital Winterthur, Winterthur, Switzerland.
[5]Division Of Haematology, University Hospital Zurich, Zurich, Switzerland.
[6]Department Of Pathology And Molecular Pathology, University Hospital Zurich, Zurich, Switzerland.
[7]Department Of Nuclear Medicine, University Hospital Zurich, Zurich, Switzerland.
[8]Institute Of Neuropathology, University Of Zurich, University Hospital Zurich, Zurich, Switzerland.
[9]Department Of Dermatology, University Hospital Linz, Linz, Austria.

Click to add notes

The clinical case



Initial Manifestation:

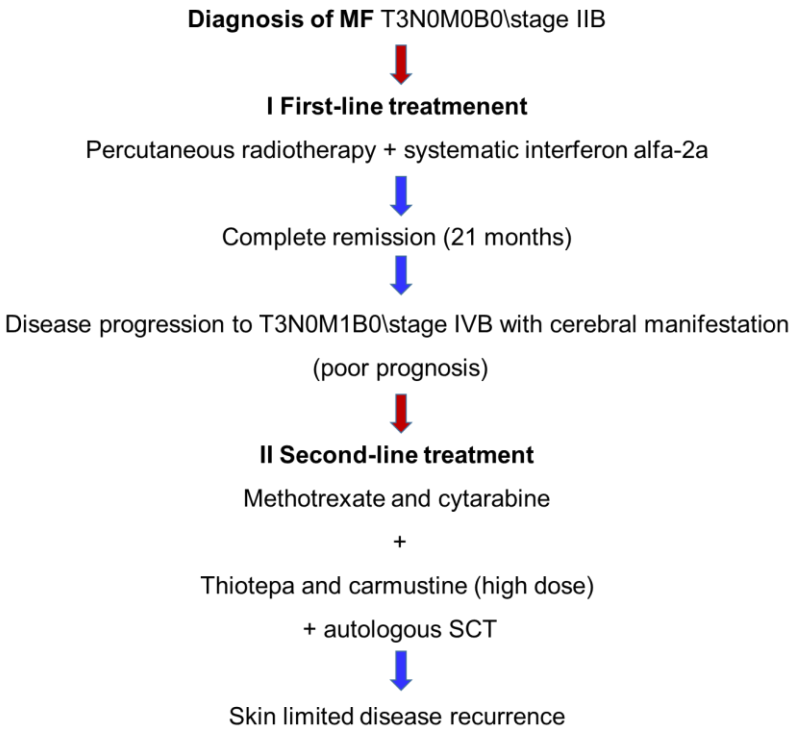
A) Clinical appearance of the skin nodule in the right mandibular region.

B) Folliculotropic mycosis fungoides (MF) with infiltration of a hair follicle by small-to-medium-sized lymphocytes without mucin deposition and some nuclear atypia (arrowheads) (haematoxylin and eosin, H&E; upper part of the image). Lymphocytes showed CD3 and predominantly CD8 positivity in the fraction of the folliculotropic lymphocytes (immunohistochemistry, lower part of image).

C) Initial clinical manifestation of MF with multiple, disseminated erythematous patches, plaques and papules.

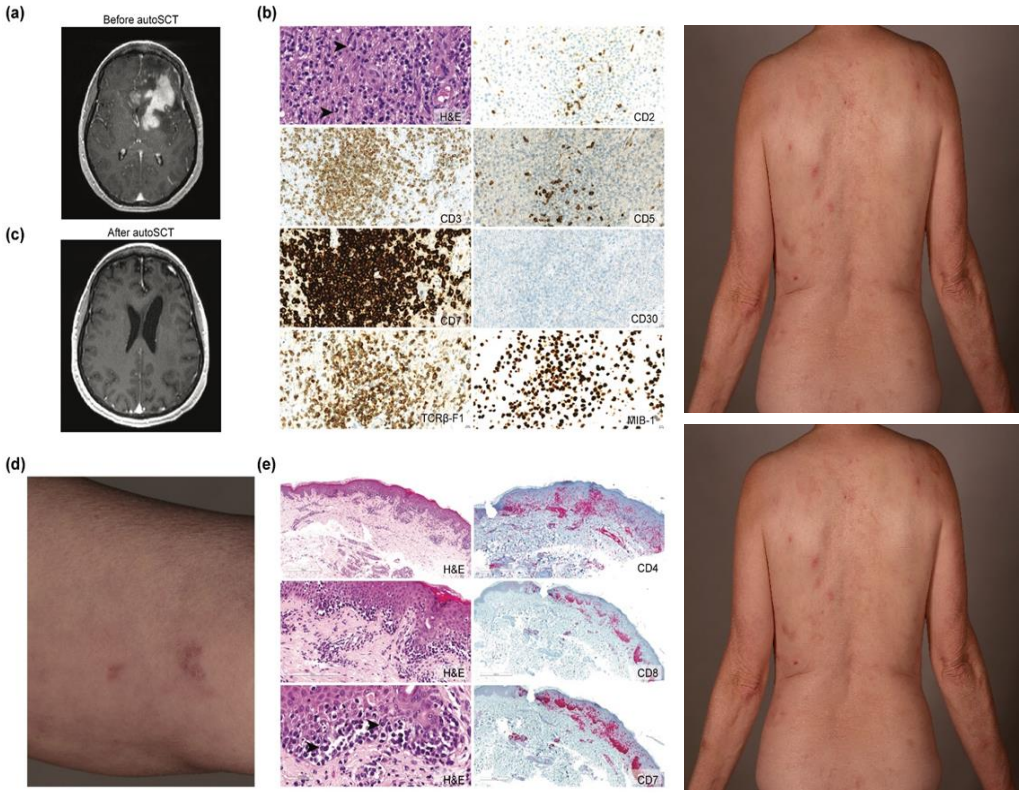
Doerschner, M. et al. *British Journal of Dermatology*, *181*(6), 1296-1302.

Treatment



Doerschner, M. et al. *British Journal of Dermatology*, *181*(6), 1296-1302.

Images before and after stem cell transplantation



A) Magnetic resonance imaging (MRI) before autologous SCT of the brain with T2 hyperintense large lesion in the left frontal lobe with involvement of the corpus callosum and basal ganglia.

B) Stereotactic brain biopsy of the frontal lobe: H&E staining demonstrates diffuse infiltration of the CNS tissue by a small round blue cell neoplasm with increased mitotic activity (yellow arrowheads) and increased apoptosis. Immunoreactivity of the neoplastic cells for CD3, CD7, TCR-beta F1 and TIA1 (not shown). Partial antigen loss for CD2 and CD5; no immunoreactivity for CD30. Proliferation index (MIB-1) >95%.

C) MRI after autologous SCT of the brain shows complete remission of the brain metastasis in the left frontal lobe.

D) Clinical appearance of recurrent lesion after auto-SCT; erythematous patch on the left upper arm.

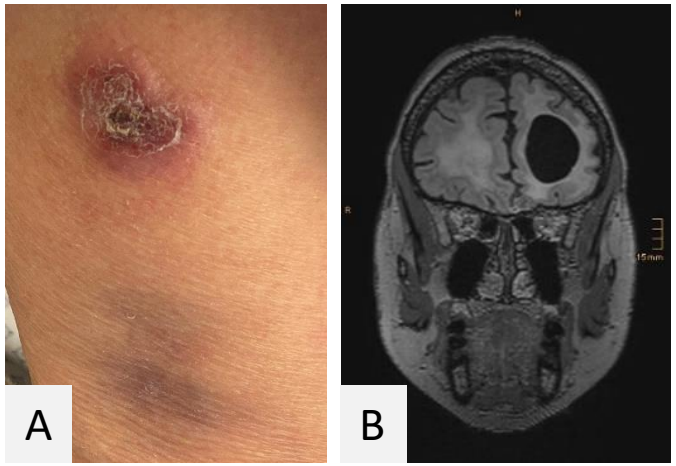
E) Skin biopsy of the recurrent lesion depicted in (D) with band-like infiltrate of lymphocytes within the superficial dermis and marked epidermotropism of atypical lymphocytes (black arrowheads). Lymphocytes showed predominantly CD8 positivity especially in the fraction of the epidermotropic lymphocytes (IHC, middle part of the image).

F) MF relapse after autoSCT.

G) Durable complete remission upon (IFN)-alfa-2a maintenance therapy.

Doerschner, M. et al. *British Journal of Dermatology*, *181*(6), 1296-1302.

Cerebral recurrence



Images upon cerebral recurrence: A) Skin tumor nodul B) cerebral MF lesion

Diagnosis of MF T3N0M0B0\stage IIB

↓

I First-line treatmenent

Percutaneous radiotherapy + systematic interferon alfa-2a

↓

Complete remission (21 months)

↓

Disease progression to T3N0M1B0\stage IVB with cerebral manifestation

(poor prognosis)

↓

II Second-line treatment

Methotrexate and cytarabine

+

Thiotepa and carmustine (high dose)

+ autologous SCT

↓

Skin limited disease recurrence

↓

III Third-line treatment

Interferon alfa-2a as a maintenance treatment after autologous SCT

Complete remission (32.5 months)

↓

Disease recurrence

↓

IV Forth-line treatment

intrathecal and intravenous administration of high-dose methotrexate and cytarabine

further progressed and exitus letalis inv 3.5 months

Cerebral involvement in MF

- Patients with plaques and tumors have greater morbidity and mortality and many develop blood and lymph nodes involvement. Ultimately many patients die due to overwhelming infections, and some develop fatal metastatic disease.
- Central nervous system (CNS) involvement is very rare and associated symptoms are scattered with poorer prognosis.
- Patient exhibited neurologic deficits as MF progressed to the CNS. Symptoms included: general systemic abnormality, vestibular, cognitive, and ocular changes

Yang, Y., & Wickless, H. (2017). *Leukemia research reports*, *8*, 14-18.